Dysphagia & Communication
Management of the ALS Patient

Becky Chapman, M.A., CCC-SLP
Roudebush VA Medical Center

Objectives

- Obtain background and review of terminology associated with ALS
- Learn early signs of ALS
- Determine next steps once early signs are noted
- Learn how to manage dysphagia in the ALS population from pre-diagnosis to end of life
- Learn methodology for choosing most appropriate augmentative/alternative communication method for each ALS patient
- Case Studies

A Little About My Background

- Began serving as primary SLP for ALS population at Indianapolis VA in 2010
- Will serve as the VISN11 Lead SLP for ALS in near future
- Current caseload of approximately 20 active ALS patients in SLP clinic
- Working with Augmentative/Alternative Communication since 2005

Amyotrophic Lateral Sclerosis (ALS) Overview

- 1 out of 10 cases of ALS are due to a genetic defect. Etiology for remaining cases is unknown.
- ALS affects approximately 5 out of every 100,000 people worldwide.
- There are no known risk factors, except for having a family member who has a hereditary form of the disease.

ALS Overview

- Average age of diagnosis is 55 years with range of 40 to 70 years.
- Men 20% more likely to develop ALS than women.
- U.S.: approximately 5,000 individuals per year receive diagnosis of ALS
- Nearly 30,000 people residing in U.S. at any time are living with ALS.
- Average time from onset to death: 3 to 5 years

ALS Overview

- Veterans are almost twice as likely to have ALS as those who have not served based on multiple Department of Defense, Department of Veterans Affairs, and Harvard studies
- Presumption of service connection within VA since 2008

http://www.alsa.org/assets/pdfs/advocacy/als_in_military_summary.pdf
What is ALS?
- Progressive neurodegenerative disease affecting nerve cells in brain and spinal cord
- Progressive degeneration of motor neurons eventually leads to their death.

What is ALS continued...
- Motor neurons die, resulting in brain’s inability to initiate and control muscle movement.
- Atrophy occurs as a result of disuse of muscles
- Affects voluntary muscle movements, including breathing

El Escorial World Federation of Neurology Criteria For the Diagnosis of ALS
- The diagnosis of ALS requires the presence of
  1) Signs of lower motor neuron (LMN) degeneration by clinical, electrophysiological or neuropathologic examination,
  2) Signs of upper motor neuron (UMN) degeneration by clinical examination, and

Diagnostic Criteria Continued
- 3) Progressive spread of signs within a region or to other regions, together with the absence of:
  - Electrophysiological evidence of other disease processes that might explain the signs of LMN and/or UMN degenerations; and
  - Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs.

LMN vs. UMN Signs
- LMN Signs:
  - Hyperreflexia
  - Spasticity
  - Extensor plantar response (up-going toes)
  - Positive jaw jerk
- UMN Signs:
  - Weakness
  - Muscle atrophy
  - Cramps
  - Fasciculations

Steps in Reaching Diagnosis of ALS
- Complete history, physical, and neuro exams for findings c/w ALS
- EEG to determine LMN degeneration
- Neuroimaging to exclude other processes
- Complete labwork to determine whether possible ALS-related syndromes are present
- Repeat clinical exam and EEG at least 6 months apart to look for progression
**Sorting out Diagnosis**  
**Terminology: Suspected ALS**
- Manifests only LMN signs in 2 or more regions  
- UMN pathology might be demonstrated at autopsy.

**Possible ALS**
- Defined on clinical grounds alone when UMN and LMN signs are in only one region or UMN signs alone are present in 2 or more regions or LMN signs are rostral to UMN signs (the latter distribution of signs needs to be differentiated from multiple non-ALS processes).

**Possible ALS Progression**
- Monomelic ALS, progressive bulbar palsy without spinal UMN and/or LMN signs, and progressive primary lateral sclerosis without spinal LMN signs and progressive primary lateral sclerosis without spinal UMN signs constitute special cases.  
- Such special cases may develop LMN or UMN signs to meet the criteria for probable ALS with time or be subsequently confirmed at autopsy by specific LMN and UMN neuropathologic findings.

**Probable ALS**
- Defined on clinical grounds alone by UMN and LMN signs in at least two regions.  
- While the regions may be different, some UMN signs must be rostral (above) the LMN signs.

**Definite ALS**
- Defined on clinical grounds alone by presence of UMN as well as LMN signs in the bulbar region and at least two of the other spinal regions  
  OR  
- Presence of UMN and LMN signs in three spinal regions.

**Definite ALS Continued**
- The important determinants of diagnosis of definite ALS in the absence of electrophysiological, neuroimaging and laboratory examinations are the presence of UMN and LMN signs together in multiple regions.

http://www.alsa.org/assets/pdfs/fyi/criteria_for_diagnosis.pdf
Clinical Features c/w ALS
1) abnormal pulmonary function test not explained by other causes,
2) abnormal speech studies not explained by other causes,
3) abnormal swallowing studies not explained by other causes,
4) abnormal larynx function studies not explained by other causes,
5) abnormal isokinetic or isometric strength test in clinically uninvolved muscles,
6) abnormal muscle biopsy with evidence of denervation.

Features INCONSISTENT w/ ALS
1) sensory dysfunction,
2) sphincter abnormalities,
3) autonomic nervous system dysfunction,
4) anterior visual pathway abnormalities,
5) movement abnormalities associated with probable Parkinson’s disease defined by DATATOP criteria,
6) cognitive abnormalities associated with clinical Alzheimer’s disease as defined by NINCDS-ADRDA criteria

Differential Diagnosis
- Brainstem Glioma
- Central Cord Syndrome
- Chronic Inflammatory Demyelinating Polyneuropathy
- Dermatomyositis/polymyositis
- Lambert-Eaton Myasthenic Syndrome
- Lyme Disease
- Multiple Sclerosis
- Posttraumatic Syringomyelia
- Dermatomyositis/polymyositis
- Sarcoidosis and Neuropathy
- Spinal Muscular Atrophy
- Primary Lateral Sclerosis

ALS vs. PLS (Primary Lateral Sclerosis)
PLS affects the upper motor neurons alone
- Significantly slower progression of symptoms
- While breathing and swallowing can be affected, typically significantly less so than ALS

When to Suspect ALS
- European Neurological Society meeting in June 2011:
  - Symptoms:
    - Clumsiness
    - Speech change, dysarthria
    - Dysphagia
    - Cramps
    - Fatigue
    - Gait changes.
  - Signs:
    - Fasciculations:
      http://www.youtube.com/watch?feature=player_detailpage&v=gZZayxWW5C5
    - Atrophy
    - Weakness

Red Flags
- “Excessive cramps”
- “Excessive fatigue”
- “Progressive tongue fasciculation”
- “Progressive weakness”

Swallowing Characteristics of ALS
- ORAL (typically affected before pharyngeal)
  - Early tongue and lip involvement
  - Increased meal duration
  - Difficulty lateralizing food and controlling food
  - To and fro movement
  - Fatigue associated with meals
  - Anterior loss of bolus
  - Increased difficulty as viscosity increases

PHARYNGEAL:
- Earlier:
  - Poor base of tongue excursion
  - Poor pharyngeal contraction
  - Delayed pharyngeal swallow initiation
- Later:
  - Reduced laryngeal elevation and excursion
  - Incomplete airway protection - penetration with subsequent aspiration
  - Possible cricopharyngeal disorder (byproduct of laryngeal issues)

Cricopharyngeal Myotomy?
- NO!
- Won't have sufficient strength with/without myotomy by the time they would need one

Modified Barium Swallow Study
- Video Examples

Dysarthria Characteristics of ALS
- Due to UMN and LMN involvement, present with mixed dysarthria
- Mixed dysarthria in ALS is combination for spastic and flaccid dysarthria
- May include:
  - Inprecise consonants (both)
  - Hypernasality (both)
  - Harsh voice (both)
  - Low pitch (spastic)
  - Reduced stress (spastic)
  - Strained-strangled voice quality (spastic)
  - Audible inspiration (flaccid)
  - Nasal emission (flaccid)
- As disease progresses, faccidity symptoms predominate

Is the ALS? What else could it be?
Comparing the dysarthria and dysphagia characteristics of ALS against other diagnoses...
Next Steps for SLPs Suspecting ALS

- Recommend Neurology consultation!!
- Comprehensively evaluate swallowing and speech, including:
  - Clinical evaluation of swallowing
  - +/- Modified Barium Swallow
  - Cranial nerve exam
  - Formal or informal motor speech evaluation
  - Evaluation for need of AAC in near future

Next Steps...

- Detailed report
- Hint at question of motor neuron disease in report
- Cease any exercises targeting strengthening muscles until diagnosis is known!

Multidisciplinary Care of the ALS Patient

**Why?**

- According to the American Academy of Neurology (2009):
  - "There is good evidence that visiting a multidisciplinary clinic can help people with ALS get the best possible care for their condition. Good evidence also shows that people with ALS who visit a multidisciplinary clinic live longer than those who don’t. There is weak evidence that people with ALS experience better quality of life by attending a multidisciplinary clinic..."


Multidisciplinary Care

According to an evidence based review in 2009 by the American Academy of Neurology:

- Two Class II studies and 1 Class III study revealed the following about patients involved in multidisciplinary clinics:
  - increased use of adaptive equipment;
  - increased utilization of riluzole, PEG, and NIV;
  - improved quality of life;
  - lengthened survival.
- However, 1 Class II study with low use of treatments found no survival benefit.

http://www.neurology.org/content/73/15/1227.full.html

Roudebush VA Medical Center ALS Multidisciplinary Clinic

- Currently meeting biweekly
- Discussion of pt needs
- Finalization of plans for clinic once space available
- Determining how best to meet VISN11 needs

Roudebush ALS Clinic

- Specialties Involved
  - Speech Pathology
  - Neurology
  - Physical Medicine & Rehab/Spinal Cord Injury MD
  - Pulmonary
  - OT/PT
  - Wheelchair Clinic
  - Social Work
  - Nutrition
Initial Patient/Family Consultation with SLP

- Topics to approach
  - Nutrition
    - Current status and barriers
    - Planning for future
  - Communication
    - Current status and barriers
    - Planning for future
    - Discuss voice banking
  - Respiration
    - Current status and barriers
    - Planning for future
  - Cognition

Determining Frequency of Follow-Up

- Based on pattern of symptoms - more frequent if bulbar
- Follow Neurology lead
- Feasibility
- Emotional needs
- Caregiver needs

ALS with Bulbar Features vs. ALS with UMN Features

<table>
<thead>
<tr>
<th></th>
<th>Bulbar Onset</th>
<th>ALS w/ UMN Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Course</td>
<td>Rapidly Progressive</td>
<td>Slow Progressive</td>
</tr>
<tr>
<td>Communication</td>
<td>Often requiring AAC</td>
<td>Slowly progressive</td>
</tr>
<tr>
<td>Swallowing</td>
<td>Early onset, progresses to inefficiency and high aspiration risk</td>
<td>Later onset and coincides with respiratory decline</td>
</tr>
<tr>
<td>Respiration</td>
<td>Difficulty with cough, hypoventilation, often requires mechanical ventilation</td>
<td>Reduced respiratory support late in disease process</td>
</tr>
<tr>
<td>Nutrition</td>
<td>High risk to fail to meet needs early on</td>
<td>Decline typically occurs in coordination with decline in ADLs</td>
</tr>
</tbody>
</table>

Management of Dysphagia in ALS

- Typically manage with diet modification and strategies
- Mealtime swallowing strategies
- Energy Conservation
- Education re: respiration's effect on swallowing
- Smaller, more frequent meals

Management of Dysphagia

- Oral supplements
- Smaller bolus volumes - sometimes
- Thicker not always better, especially with progression
- Regular f/u and repeat clinical exam +/- MBS
- Consideration of PEG sooner than later

Management of Communication in ALS
### Five Stages of Speech Decline in ALS (Mathy, et. al.)

**Stage 1**
- **No Detectable Speech Disorder**
- Diagnosis has been made, but often speakers do not yet exhibit speech symptoms in those with spinal presentation.
- Listeners: note no difference in rate, precision, or loudness

**Stage 2**
- **Obvious Speech Disorder with Intelligible Speech**
- Both the speaker and listener notice changes in speech - speakers may perceive extra effort needed for speech.
- Worse with fatigue or stress
- Speakers may begin to compensate:
  - Decreased speaking rate
  - Decreased length of breath groups

**Stage 3**
- **Reduction in Speech Intelligibility**
- Changes in speaking rate, articulation, and resonance are all evident.
- Speakers can/do modify production
- Strategies:
  - Slow speaking rate
  - Conserving energy
  - Increasing precision
  - Communication breakdown plan

**Stage 4**
- **Natural Speech – Supplemented w/ AAC**
- Natural speech no longer functional means of communication in all situations.
- Strategies/interventions:
  - Alphabet supplementation
  - Alerting signal for gaining attention
  - Augmented telephone communication
  - Portable writing systems

**Stage 5**
- **No Functional Speech**
- Speakers with advance bulbar ALS have lost functional speech due to profound weakness.
- May vocalize for emotional expression or w/ extreme effort
- Strategies/intervention:
  - Establishing reliable yes/no
  - Eye-gaze systems
  - Integrated, multipurpose AAC systems

### Maintaining Use of Natural Speech (David Beukelman)
- **Behavioral Interventions**
- **Environmental Interventions**
- **Prosthodontic Interventions**
- **Supplemented Speech Interventions**
**Behavioral Interventions**
- Speaking rate modification
- Maintain coordinated respiratory patterns
- Reduce fatigue
  - Conserve energy for communication
- Eliminate oral or non-speech exercises

**Prosthodontic Interventions**
- Palatal lift (controversial and temporary)
- Palatal augmentation (drop-down)
- Voice amplification (i.e., Spokeman, Chattervox)

**Environmental Interventions**
- Optimize hearing of frequent listeners
- Optimize adverse speaking situations
  - Reduce background noise
  - Mute TV
  - Amplify speaker in meetings, groups, & noise
  - “Private conference room”

**Supplemental Speech Interventions**
- Topic boards
- Alphabet boards

**Low Tech AAC Options**
- Handwriting: Boogie Board, dry-erase board
- Partner-assisted auditory scanning
- Partner-assisted manual scanning
- Light-technology optical pointing with head mounted laser
- Facial expression
- Yes/no questions

**High Tech AAC**
- Determining Candidacy:
  - Familiarity with computers
  - Attitudes towards high tech AAC
  - Support system
  - Extremity function
  - Cognitive function
  - Vision (premorbid, not related to ALS)
  - Language
Determining Candidacy

- Ambulatory Status
- Size limitations of home
- Respiratory function
- Anticipated progression of disease
- Is this device overkill?
- What other needs will this device meet?
- Is there a partner to participate in lower tech communication as back-up?

Speech Applications - 100 to choose from!

- Proloquo2Go
- TalkTablet
- Verbally
- SpeakIt
- Predictable
- Phrase Board

Cognition and ALS

- Why is cognition affected?
  - Dementia as chance-co-occurrence
  - Unlikely given lack of typical Alzheimer’s brain characteristics in pts with ALS and dementia in some cases.
  - Multisystem involvement, including frontal lobes
  - Sasaki et al. studied multisystem involvement in pts who survived beyond point of respiratory failure. ALS progressed to include:
    - Putaminal atrophy
    - Cerebral cortex
    - Dentate and red nuclei
    - Thalamus
    - Mamillary body

High Tech AAC Options

- Dedicated Systems with eye gaze capability
- Tobii AT1
- Dynavox
- LC Technologies
- iPad with speech applications

Cognition and ALS

- Classic view of cognition being unaffected is being challenged:
  - According to studies by Massman et al., as cited in Beukelman, >1/3 patients with ALS displayed clinically significant impairment (at or below 5th percentile on at least 2 of 6 neuropsychological tests.
  - Cognitive problems more associated with bulbar onset
  - Some believe Dementia with ALS should be classified as one of the frontal lobe degenerative dementias

References